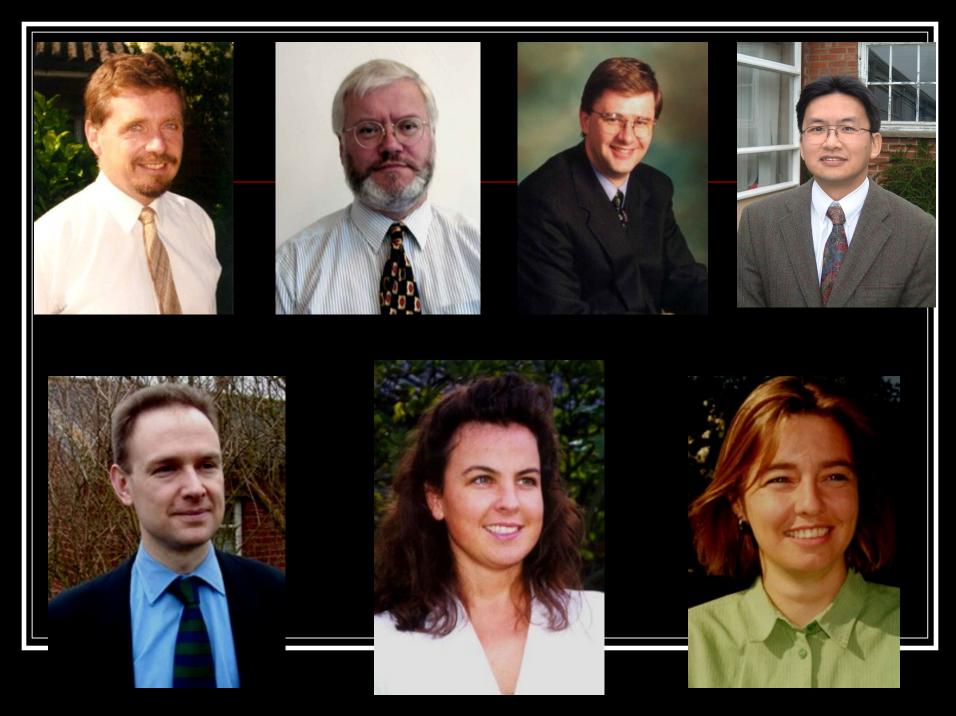
Lesley Bennett Consultant Physician

Oxford Centre for Respiratory Medicine Churchill Hospital

Respiratory Services

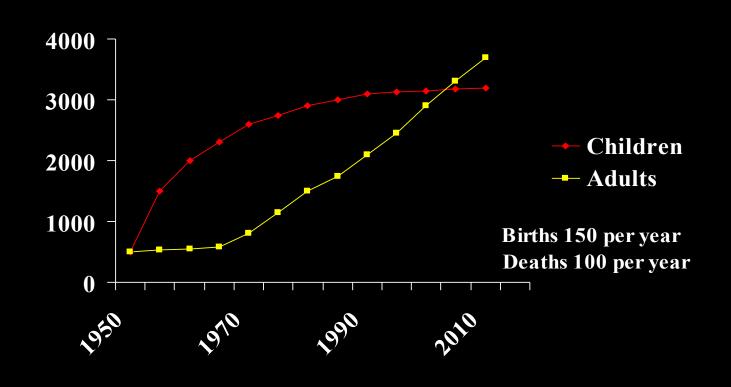
- 5 WTE Consultants
- Inpatient care of 20 beds on the respiratory unit including 2 high dependency beds
- Approx 11000 out-patients per year
- Treatment centre
 - 1200 day cases
 - Rapid assessment for urgent patients



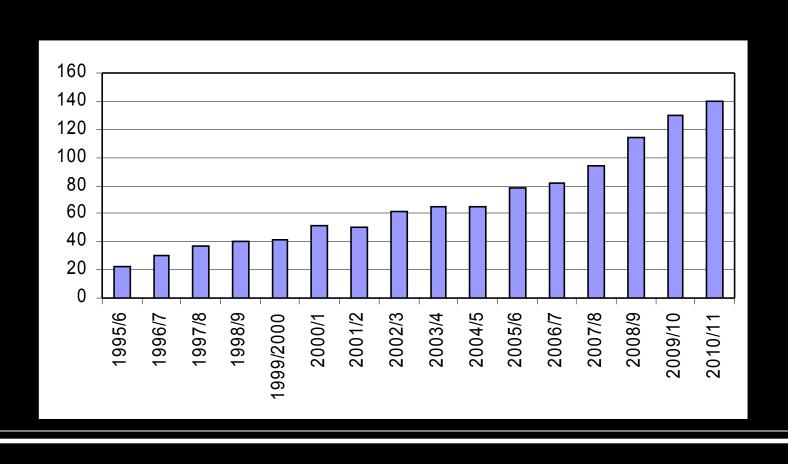
Specialist clinics/services:

- Sleep related breathing disorders including nasal CPAP and non-invasive ventilation
- Cystic fibrosis (only adult centre in Thames Valley SHA)
- Pleural disease including diagnostic thoracoscopy
- Interventional bronchoscopy (endobronchial palliation)
- COPD clinic
- Bronchiectasis clinic
- Sarcoidosis/ILD clinic

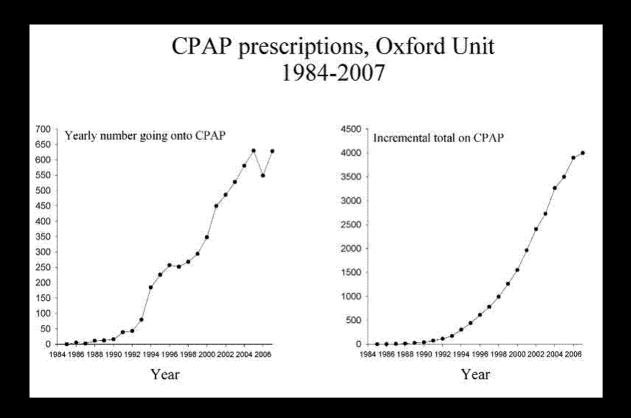
Epidemiology - England & Wales



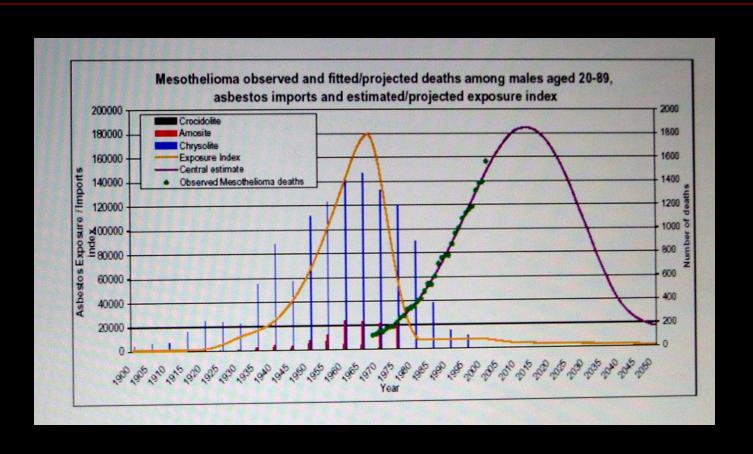
Predicted growth in Oxford adult cystic fibrosis patients



Sleep apnoea

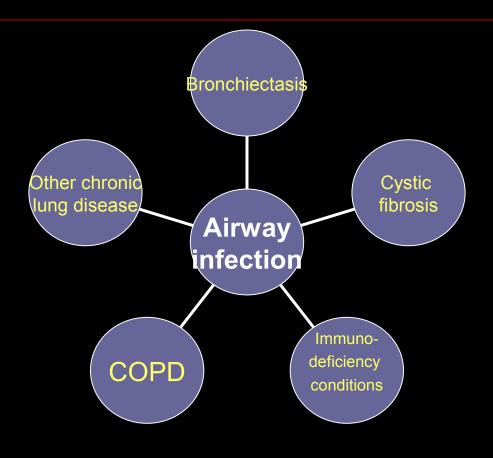


Malignant mesothelioma



Airway infection in chronic lung disease

Associated conditions



Bronchiectasis

- Abnormal and permanent distortion bronchi
- Congenital (CF, immune deficiency, PCD) or acquired (previous pneumonia, TB etc.)
- Adults, mostly 'idiopathic'



Incidence

- Largely unknown
- Apparent 'increase' in incidence probably due to more cases being diagnosed with HRCT
- Bronchiectasis has been identified using HRCT in up to 15%-30% of patients diagnosed in primary care with chronic bronchitis and COPD

Pathophysiology

Infection

Impairment of host defense/drainage

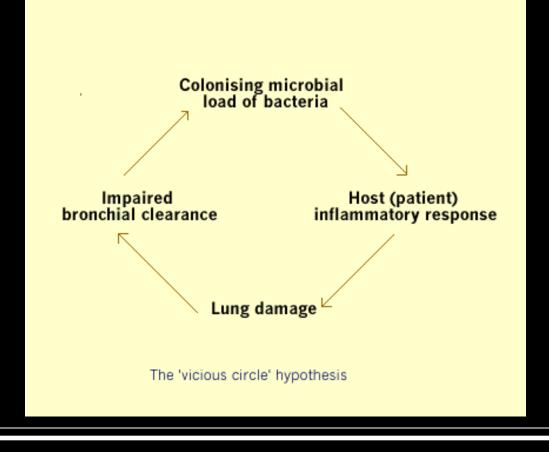
Inflammatory host response (damage to bronchial wall)

Abnormal bronchial dilatation with bronchial wall destruction

Impairment of clearance of secretions

Colonization and infection with pathogenic organisms,

Vicious cycle of infection/lung damage



Clinical features that might suggest bronchiectasis:

- Cough and daily mucopurulent sputum (75%)
- Dyspnoea (72%)
- Obstructive spirometry without significant smoking history
- History of frequent episodes of 'bronchitis'
- Pleuritic chest pain (intermittently in 40%). Usually secondary to chronic coughing, occasionally acute exacerbation.
- Other: haemoptysis, weight loss (severe bronchiectasis)

Examination

- Crackles may/may not be helpful
- FEV1/FVC useful, for documenting severity and tracking changes

Systemic markers of infection often not present

Diagnosis

- Clinical history
- CXR can be normal
- Characteristic findings on CT scans
- Often airflow obstruction on spirometry

Natural History

- Incurable
- Active management reduces symptoms and delays/prevents further lung damage
- Complications include lung abscess, haemoptysis, empyema, pneumothorax
- Currently, mortality is related more to progressive respiratory failure and cor pulmonale than to uncontrolled infection

Outcome

- Few studies on outcome
- Approx 1 in 5 will have premature death (mean age 53 years)

Treatment

- Disease education/self-management plan
- Regular physiotherapy
- Prompt antibiotics for infection
- Multi-disciplinary clinic
- Telephone advise service

Education

Patients need to understand risk of progression and 'vicious circle'

Agree what is 'normal symptoms' and therefore what is acceptable and clarify the aims of treatment

Determine 'normal symptoms'

- No symptoms
- Recurrent acute bronchitis (infective exacerbations) with no background symptoms
- Recurrent acute bronchitis on a background of daily mucoid sputum
- Daily purulent sputum without systemic/generalized symptoms plus/minus recurrent acute bronchitis
- Daily purulent sputum with systemic/generalized symptoms plus/minus recurrent acute

Mild

Severe

Agree aims of treatment

Ideal

Realistic

Background symptoms

Abolition

Minimise

Acute episodes of bronchitis

Prevention

Reduce severity

Disease progression

Prevention

Prevention

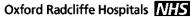
Adverse effects

None

Minimise

Disease education

- Be aware of changes in sputum (viscosity, colour, volume)
- Self-management plan
 - Sputum specimen
 - Initiate 'stand-by' antibiotics
 - Contact service if no improvement in 3-4 days



SELF MANAGEMENT PLAN FOR **BRONCHIECTASIS PATIENTS**

The doctor and physiotherapists will have talked to you about the importance of prompt treatment for infective exacerbations. It is therefore important to be aware of symptoms of an exacerbation as follows:

- An increase on the volume of your sputum from your normal day-to-day amount
- A change in colour of your sputum
- Feeling more tired and generally not well
- Your chest feeling constricted or congested
- Evidence of soreness or pain in your chest
- Shortness of breath, and wheeze

If you have more than two of the above symptoms, and do not know what to do, please contact the bronchiectasis team on 01865 225713 or 01865741841 asking for bleep 1234 for advice (Monday to Friday, 9:30 to 12:30)

- If possible send a sputum sample to your local GP surgery or by post to the laboratory at the John Radcliffe Hospital, using the correct postal packaging provided by the hospital.
- . If the sample has been sent to the John Radcliffe Hospital. Please contact the bronchiectasis team for the result after four
- Start your prescribed reserve course of antibiotics.
- . If you are not responding to the treatment within three to four days, contact the bronchiectasis team.
- If you are having recurrent infections i.e monthly, please contact the bronchiectasis team to discuss if you need an earlier review.

Antibiotics

- Recommend high doses for at least 2 weeks
- In vitro sensitivity not necessarily representative of in vivo action
- Treat according to organism from most recent sputum microbiology
- H Influenzae most common in mild/mod disease
- Pseudomonas aeruginosa more common as disease progresses

Which antibiotic?

Amoxicillin 500mg tds for 2 weeks (clarithromycin 500mg bd)

Second-line

Co-amoxiclay 625 tds for 2 weeks

One that has worked before!

Long-term antibiotics

- Desperate measure!
- In selected patients only
- In response to recent change in symptoms
- Usually rotate every 28 days
- Only use if oral antibiotics work

Pseudomonas aeruginosa

- Chronic infection usually associated with significant decline
- Once established, difficult to clear
- Treatment of exacerbations much more complicated
- Aggressive management of PsA infection, 5 year survival increased from 54% to 82% in CF

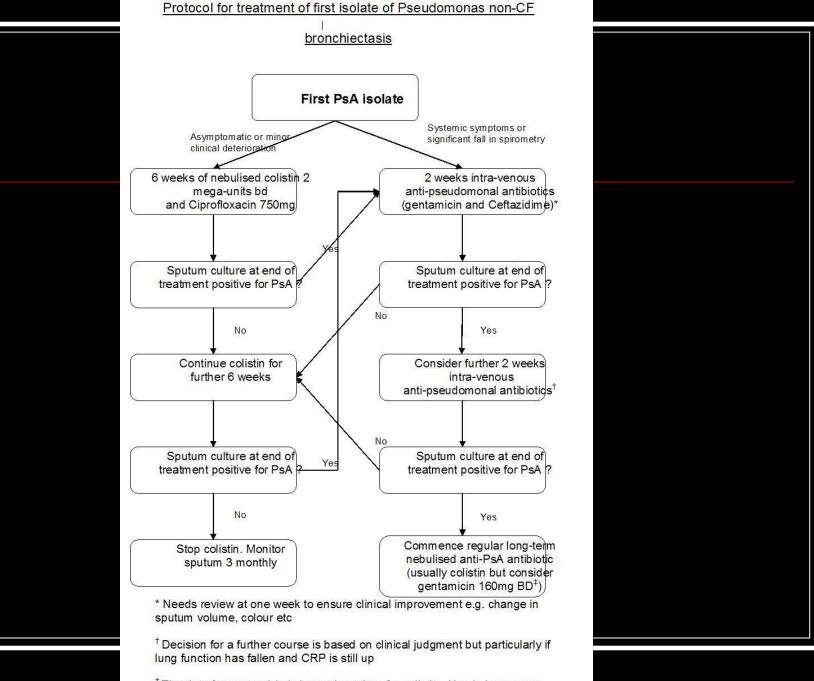


Pseudomonas aeruginosa – first isolate

- Colistin nebulised 2 mega-units bd
- Ciprofloxacin 750mg
- 6 weeks

□ Valerius et al (1991)

- If still colonised at 6 weeks
- Check compliance with colistin
- Consider 2 weeks intra-venous anti-pseudomonal antibiotics



[‡] The data for gentamicin is less robust than for colistin. Use in less severe cases and monitor progress. In the first 6 months if spirometry falls or requires an additional course of iv antibiotics, switch to nebulised colisitin

Chronic infection with PsA

Usually require iv antibiotics for exacerbations

Some patients suitable for selfadministration of home iv antibiotics

Long term nebulised antibiotics reduce frequency of exacerbations

Physiotherapy

- Aims are to reduce airway obstruction by improving the clearance of secretions
- To reduce the severity of the infection by clearing infected material
- Variety of techniques according to technique/disease severity
- Exercise

Physiotherapy

- Active methods preferred
- E.g. ACBT, AD

Postural drainage and percussion used much less in adults

Flutter device

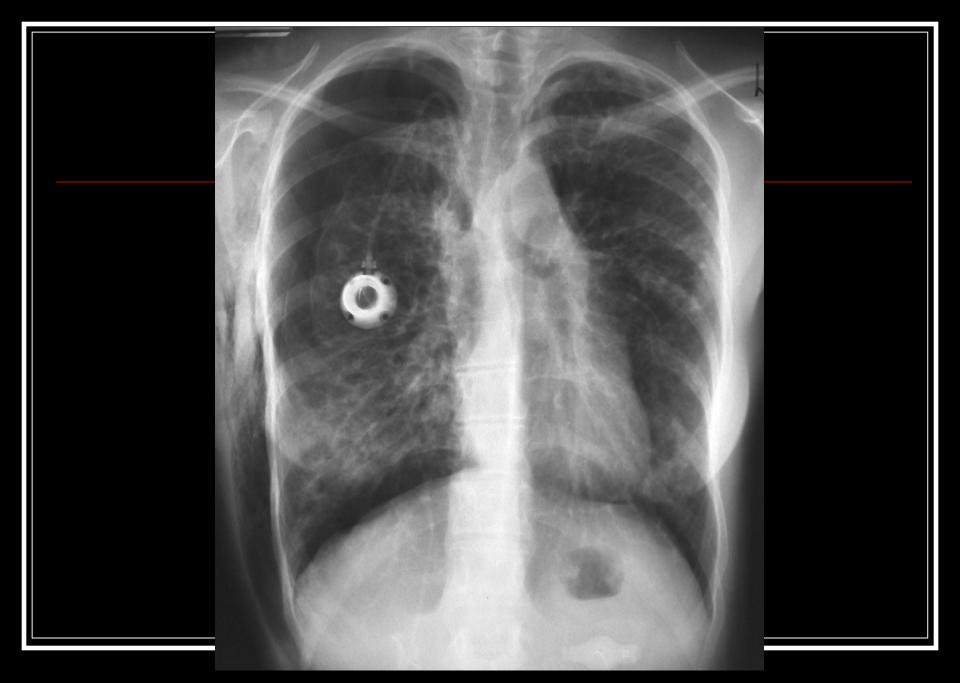


Other treatments

- Immunisation
- No role for inhaled steroids (unless co-existing asthma)
- Consider bronchodilators
- Anti-inflammatory drugs no role
- Nebulised antibiotics (usually for Pseudomonas)
- Surgery very rarely appropriate
- Transplant

Total implantable venous access devices (portacath)





Case

54 year old woman

Recurrent episodes of bronchitis

Now some breathlessness

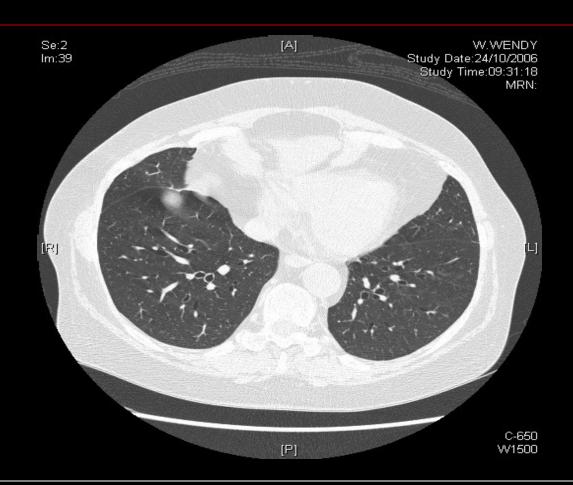
Smoked until age 30, stopped 'smokers cough'

Childhood whooping cough

Investigations

- CXR Normal
- Spirometry
 - FEV1: 1.8 (predicted 2.4)
 - FVC: 2.8 (predicted 2.9)
 - FEV1/FVC = 64%
- Sputum MC and S H Influenzae
- HRCT thorax

HRCT



Investigations

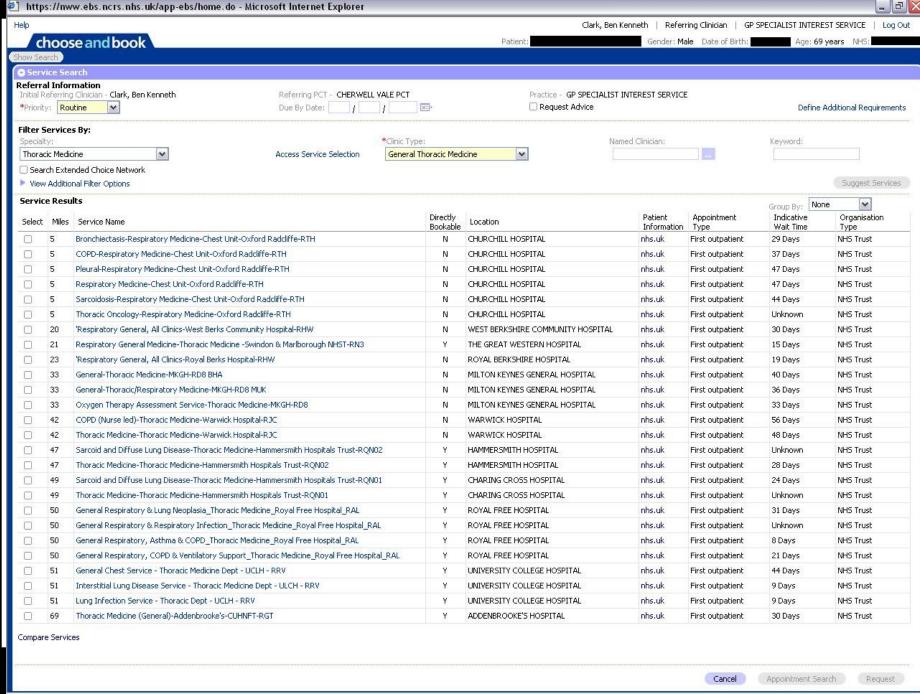
- Immunoglobulins Low IgG
- Aspergillus RAST

Common Variable Immunodeficiency

Other tests only if clinical suspicion (CF genotype/sweat test/ciliary studies)

Bronchiectasis service

- Patient telephone support service
 - contact point for the bronchiectasis patients/GPs.
 - Medical assessment can be arranged for those who need it.
- Annual review appointment (Instead of 3-4 monthly review)
 - For stable patients who have self-management plan
 - Disease education
 - Microbiological surveillance
 - Monitoring disease progression
- Home intravenous antibiotics
 - Commenced in hospital (but 2-3 day stay rather than 14 day).



Chest Radiograph, Pulmonary Function Tests, Reversability Testing, Arterial Blood Gases, Capillary Blood Gases, Heaf Test, Mantoux, Fine Needle Aspiration, FNA, Pleural Aspiration, Skin Prick Testing, Flight Assessment

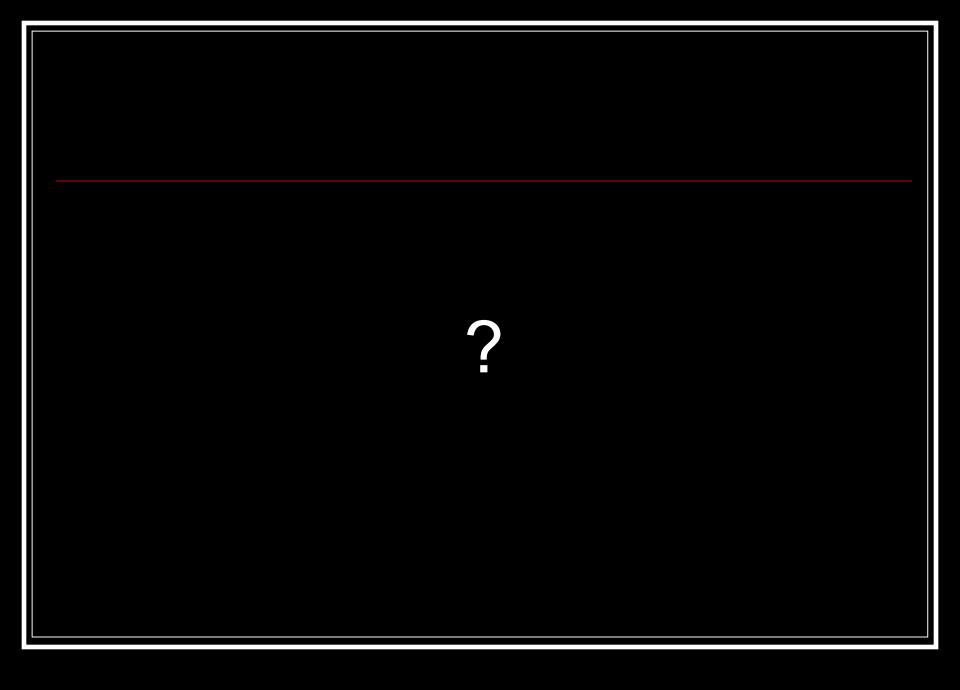
Service Notes

This is a MDT Clinic with a specialist Nurse and Respiratory Physiotherapist

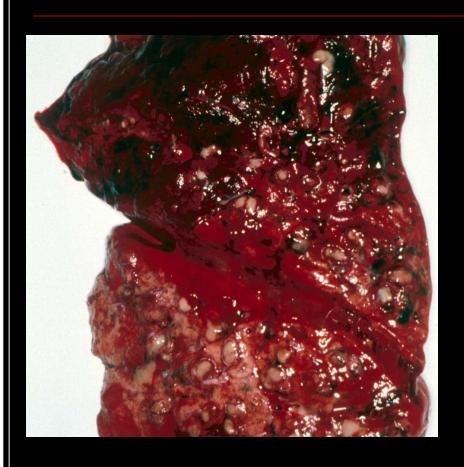
Booking Details

Booking Telephone Number: 0845 3308888 Hours of Operation: 8am and 9pm; Monday - Friday; 9am and 5pm; Saturday - Sunday

Please contact our Patients Appointments Bureau, which is open 7 days a week.



Cystic Fibrosis – an adult disease



Cystic Fibrosis (CF) is the UK's most common, life-threatening, inherited disease

Cystic Fibrosis affects over 7,500 people in the UK

Failure of CFTR leads to the main clinical problems in CF due to increased viscosity of mucus in lungs and pancreas

Cystic fibrosis survival

- Dramatically improved survival in last 30 years due to:
 - maximising nutrition using enzymes, supplements and enteral feeding
 - Aggressive management of pulmonary disease
 - Lung transplant
- Average life expectancy is 31 years

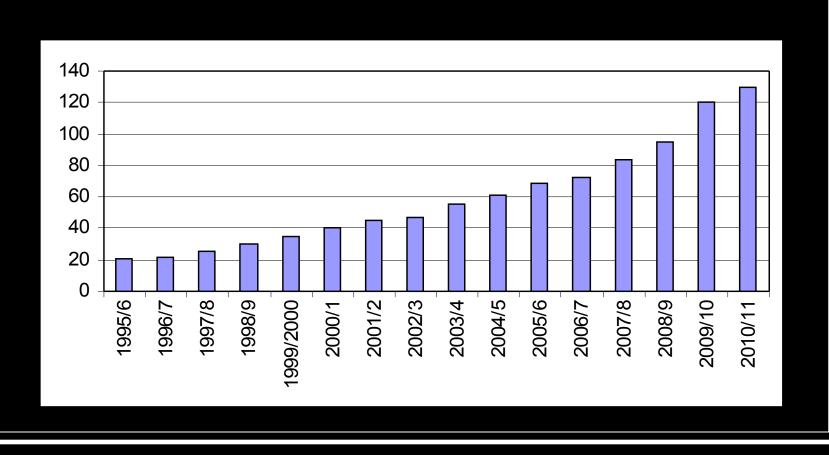
Changing epidemiology

Percentage of CF patients surviving to age 5

Year born	Males	Females
1968-70	75%	73%
1977-79	88%	87%
1989-1991	97%	97%

- More patients are surviving to adulthood
- 50% of CF patients are now 16 or over and 30% are over 20

Predicted growth



Treatment – Lung disease

- Daily physiotherapy
- Frequent antibiotics
- Intra-venous antibiotics
- Inhalers
- Daily nebulised antibiotics
- Oxygen
- Non-invasive ventilation
- Lung transplant

DISEASE PROGRESSION

Other treatment

Other complications

- Daily enzymes
- Nutritional supplements/NG or PEG feeding
- Vitamin supplements
- Insulin

- Cystic Fibrosis Related Diabetes mellitus
- Bowel Obstruction
- Gastro-oesophageal reflux
- Pancreatitis
- Liver and biliary disease
- Osteoporosis
- Nasal polyps and sinusitis
- Arthropathy, arthritis and vasculitis
- Infertility
- Pneumothorax
- Haemoptysis

Tertiary service

- Multi-disciplinary specialist team
 - 2 consultants
 - 2 physiotherapists
 - 3 CF specialist nurses
 - Dietician
 - Pharmacist
 - Psychologist
- Open access to service
- Liaison with other services:
 - Joint diabetic and hepatology clinics
 - Insertion of Intra-vascular devices
 - ENT
 - Interventional Radiologist
 - Oxford Fertility Unit
 - Silver Star Service
 - GI Surgeons

Disease progression

- Deterioration with increasing age
 - Increasing disability
 - Frequent hospital admissions/attendances
 - Respiratory failure
 - Diabetes
- Expensive to treat
 - Banding system

'Banding' of CF costs

Band		2006/07
1	Predominantly out-patient treatment	£2313
П	Intravenous antibiotics 3-4 times a year	£9663
Ш	Intravenous antibiotics 3-4 times a year, some as inpatient	£29666
IV	Severe disease. Intravenous antibiotics 3-4 times a year predominantly as in-patient. Gastrostomy. Diabetes.	£63245
V	Frequent hospital admission. On non-invasive ventilation. Transplant assessment	£99632

Predicted Costs

Year	2006/ 2007	2007/ 2008	2008/ 2009	2009/ 2010	2010/ 2011
Band					
I	£60138	£67704	£85884	£102960	£115878
II	£347868	£393822	£485392	£595458	£668334
Ш	£355992	£403013	£518336	£609372	£707540
IV	£189735	£198273	£207195	£288692	£301684
\mathbf{V}	£498160	£624696	£761607	£909576	£950504
T Total	£498160 £1,950,053	£624696 £2,312,204	£761607 £2,820,021	£909576 £3,415,634	£950504 £3,694,444



